Case Report - Developmental Disorder

Midline segmental odontomaxillary dysplasia

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ABSTRACT

Segmental odontomaxillary dysplasia (SOD) is a rare developmental disorder of the maxilla, characterized by variability of its clinical and radiological features and may mimic other fibro-osseous lesions. Clinically, the disorder is often diagnosed in early childhood due to a unilateral buccolingual expansion of the posterior alveolar process, gingival enlargement, absence of one or both premolars in the affected region, delayed eruption of the adjacent teeth, and malformations of the primary molars. We describe a rare case of a SOD in a 19-year-old female comprising findings similar to earlier reports, but for the first time SOD is reported along midline. She presented with pre-maxillary enlargement and abnormal pattern of eruption of anterior maxillary permanent teeth. Radiographic imaging showed abnormal bony trabeculation. Histopathologic findings showed characteristic features of SOD. We herein report a case of this rare unusual anomaly and review the literature. Clinicians should be aware of its presence when encountering patients presenting with facial asymmetry unresponsive to treatment.

Keywords: Hemimaxillofacial dysplasia, segmental odontomaxillary dysplasia, facial asymmetry, maxillofacial developmental anomaly

INTRODUCTION

Segmental odontomaxillary dysplasia (SOD) has been delineated from other bone and odontogenic dysplasias, and only recently has been described as a separate entity.[1,2] This condition was first introduced as hemimaxillofacial dysplasia (HD) by Miles *et al.* in 1987.[1] SOD is characterized by unilateral maxillary enlargement, gingival hyperplasia, facial asymmetry, ipsilateral dental abnormalities, and an unusual radiographic bone pattern.[2] Immature woven bone forming irregular patterns are seen histologically. To date, less than 40 cases have been reported in the English literature. All cases reported appear to represent sporadic occurrence with no inheritance pattern. The condition commonly diagnosed in the first decade.[1-12] Most of the times, clinicians have misinterpreted the condition as hereditary gingival fibromatosis because of the maxillary enlargement and thickened gingivae. The poor-quality bone encountered on biopsy may have suggested atypical fibrous dysplasia. We here report the unique case of SOD, presenting with pre-maxillary enlargement and abnormal pattern of eruption of anterior maxillary permanent teeth.

CASE REPORT

A 19-year-old female complained of an asymptomatic expansion of the anterior maxilla in midline. At age 9, the parents had sought care from a pediatric dentist because the permanent incisors in the child's maxilla failed to erupt. The gingival tissue over the unerupted permanent incisors was described as excessive and fibrous and was removed surgically to permit permanent incisors eruption. Her family history was unremarkable.

On clinical examination premaxillary alveolus was markedly expanded, with several protuberances visible on the labial aspect. The degree of expansion was greater than usual at age 9. All the four permanent incisors and both canines were somewhat malpositioned with wide diastemas. The enamel of the permanent incisors was hypoplastic, appearing yellowish. The marginal gingiva especially on the buccal aspect of the anterior maxillary permanent teeth appeared somewhat thickened. No pain was elicited when the area was digitally palpated [Figure 1].

Periapical and panoramic radiographs and CT scan revealed an ill-defined, coarse, irregular trabecular bone pattern superimposed over the anterior maxillary permanent teeth [Figure 2].

Surgical recontouring of anterior maxilla together with extraction of maxillary anterior permanent teeth was performed under general anesthesia [Figure 3].

Histologic examination revealed fragments and trabeculae of immature woven bone forming irregular pattern. Mature lamellar bone was not present [Figure 4]. A final diagnosis of SOD was made based on the correlation of the clinical, radiographic, and microscopic findings.

Patient was clinically asymptomatic after 6-month follow-up and was placed on interim prosthetic rehabilitation.

DISCUSSION

SOD is an unusual unilateral maxillary developmental anomaly with characteristic clinical, radiographic and histologic findings. Since its original description as hemimaxillofacial dysplasia by Miles and colleagues[1] less than 40 cases have been reported in the English literature.

Etiopathogenesis

The etiopathogenesis of SOD is unknown, although it is suggested that it might be due to a localized developmental abnormality *in utero*,[2] a unilateral developmental field defect involving anlagen of the first and second branchial arches,[5] or a postzygotic mutation or a similar phenomenon affecting a progenitor cell and could have resulted in an altered clone of cells that influenced the morphogenesis of the ecto-mesodermal tissues in a segment of the maxilla and the overlying facial tissues.[8]

Figure 1: Extra-oral and intra-oral clinical presentations showing markedly expanded pre-maxillary alveolus, with several protuberances visible on the labial aspect. All the four permanent incisors and both canines were somewhat malpositioned with wide diastemas

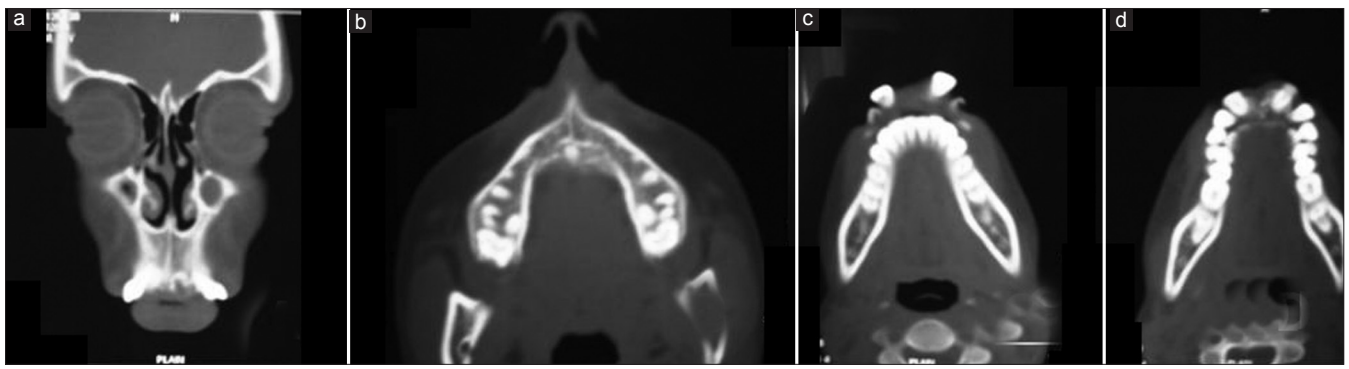

Figure 2: CT scan revealing an ill-defined, coarse, irregular trabecular bone pattern superimposed over the anterior maxillary permanent teeth

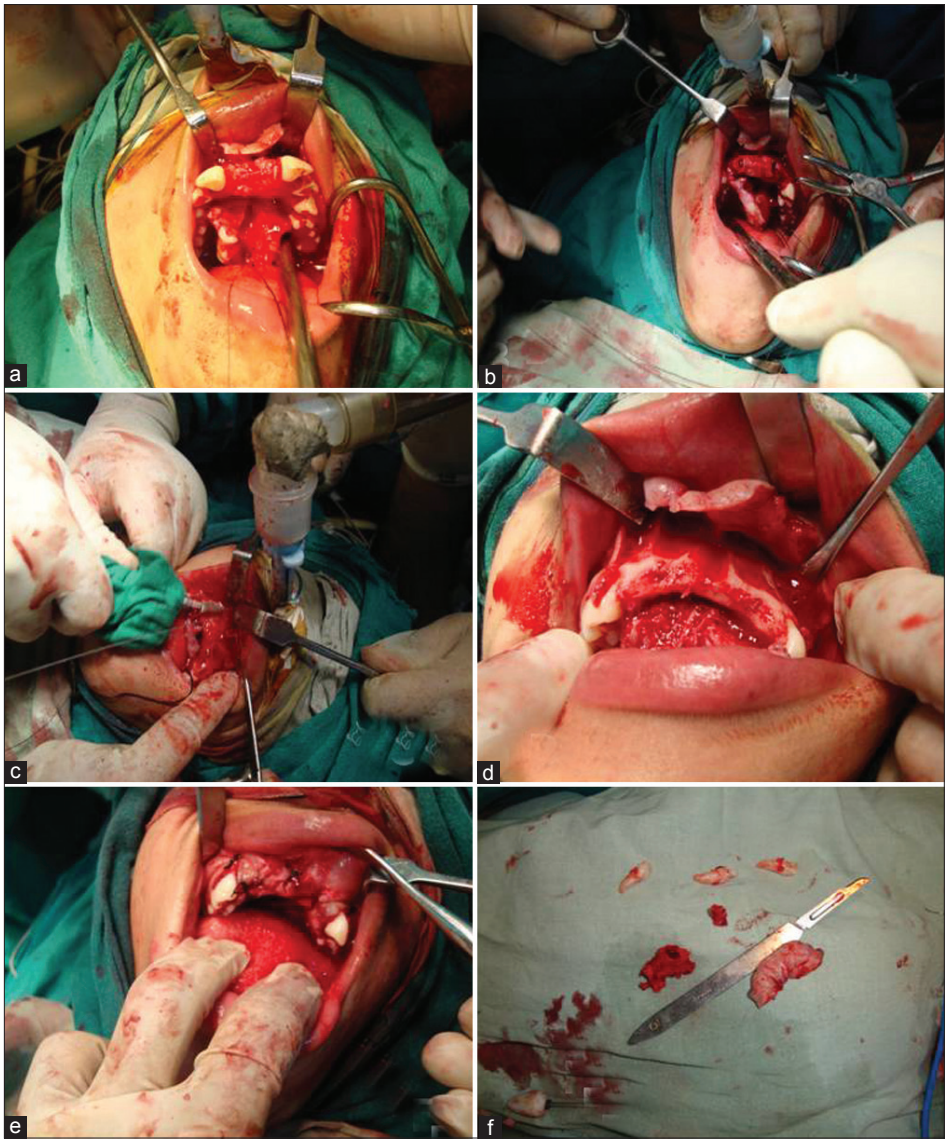

Figure 3: Surgical recontouring of anterior maxilla together with extraction of maxillary anterior permanent teeth

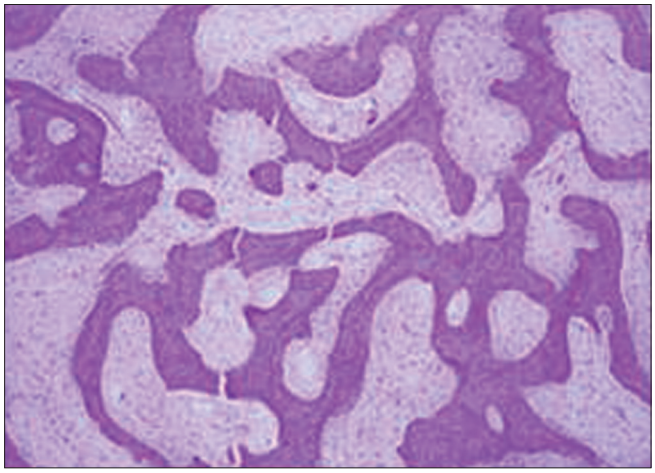

Figure 4: Photomicrograph showing uninflamed fibrous stroma and irregular trabeculae of woven bone. There is no osteoblastic rimming of the bone trabeculae (H & E stain, 20x)

A viral or bacterial infection of the maxillary division branches of the trigeminal nerve have been proposed as initial causative factors in SOD.[3]

Epidemiology

All cases reported appear to represent sporadic occurrence with no inheritance pattern. Males were affected more than females with a male-to-female ratio of 1.8:1. The condition is commonly diagnosed in the first decade. The right and left sides of the maxilla are almost equally affected.[1-12] However, in present case, female patient presented with pre-maxillary alveolus was markedly expanded noticed first at the age of 9, which is first of its kind reported in world English literature.

Clinical and radiographic features

Clinically, SOD usually presents as a non-progressive facial asymmetry, ipsilateral gingivo-dento-alveolar maxillary involvement from the canine eminence anteriorly until the

maxillary tuberosity posteriorly, and with or without facial cutaneous lesions. There are a number of reported dental abnormalities such as enamel defects, absence of one or more permanent teeth, and distal displacement of permanent molars that may require consultation with an oral surgeon and orthodontist. In present case, pre-maxillary alveolus was markedly expanded (first of its kind reported in world English literature), with several protuberances visible on the labial aspect. The degree of expansion was reportedly greater than at age 9. All the four permanent incisors and both canines were somewhat malpositioned with wide diastemas. The marginal gingiva especially on the buccal aspect of the anterior maxillary permanent teeth appeared somewhat thickened.

Radiographically, SOD shows mainly bony sclerosis with thickened and coarse irregular bone trabeculation pattern producing an ill-defined opacity.[2,3,6,7] Similar presentations were seen in present case also.

Clinical and radiological features of different grades of SOD are summarized in Table 1[13] as:

Facial cutaneous manifestations of SOD are variable and affect up to 48% of SOD patients of whom 80% are males.[1–12] The most common facial cutaneous manifestations are hypertrichosis followed by facial erythema, lip hypopigmentation, and Becker's nevus.[1,3,5,6,8,12] Other cutaneous manifestations include "hairy nevus," discontinuity of the vermilion border, depression of the cheek, and facial hyperpigmentation.[5,7,12] When hypertrichosis is the associated cutaneous lesion, it is only limited to males.[1,5,8,12]

Classification

Othman *et al.*[13] have recognized SOD into two major types:

1. Type I classical with gingivo-dento-alveolar manifestations without extraoral facial involvement
2. Type II cutaneous with dermato-gingivo-dento-alveolar involvement. Type II is further differentiated into four subtypes:
   - Type IIa hypertrichotic,
   - Type IIb pigmentory,
   - Type IIc erythematous, and
   - Type IId commissural defect.

Furthermore, combinations between Type II subtypes are possible such as Type IIab with hypertrichotic–pigmentory combination, Type II abd with hypertrichotic–pigmentory–commissural defect combination, and Type II bc in our patient with pigmentory–erythematous combination.

Table 1: Clinical and radiological features of SOD[13]

75%–100%	50%–75%	25%–50%	<25%
Mild-to-moderate facial asymmetry	Non-progressive growth	Cutaneous facial lesion	Absent root development
Buccolingual alveolar and gingival thickening	Diagnosis in first decade	Splayed roots	
Hypodontia	Affected male	Reduced size of pulp chambers	
Widely spaced/ displaced teeth	Hypoplastic teeth	Large pulp chambers	
Ill-defined radiopacity	Increased mesiodistal crown dimension		
Vertically oriented bone trabeculation	Delayed eruption/ impaction of teeth		
Thickened bone trabeculation	Root resorption/ill-defined root morphology		
Reduced size of ipsilateral maxillary sinus			

Histopathology

Histologic characteristics of the anomaly are non-specific, non-inflammatory connective tissue thickening and fibrosis of gingival tissues.[1,2] Alveolar bone shows absence of mature lamellar bone, presence of thick trabeculae of immature woven bone with prominent resting/reversal lines, uninflamed fibrous stroma, and lack of osteoblastic and osteoclastic activity.[1-4,6] Dental defects include circum-pulpal dentin tubular defects, irregular pulp/dentin interface with pseudo-inclusions, focal deficiency in odontoblastic layer, and coarse fibrous pulp with pulp stones.[11] Present case presented with similar histological findings.

Diagnosis

SOD may go unrecognized or misdiagnosed.[6,12] Differential diagnosis includes monostotic fibrous dysplasia, regional odontodysplasia, gingival fibromatosis, segmental hemifacial hypertrophy, and various benign odontogenic tumors.[3,4,6,12] In present case, final diagnosis of SOD was made based on the correlation of the clinical, radiographic, and microscopic findings.

Clinical and radiological similarities between SOD and these lesions exist, which makes histopathologic examination necessary to accurately establish the diagnosis and exclude other pathological entities.

Differential diagnosis

Monostotic fibrous dysplasia

Fibrous dysplasia or a related fibro-osseous lesion are to be considered because of the midline maxillary expansion and the atypical bone pattern noted on radiographic examination.[14] In fibrous dysplasia the teeth in the area are normal. In large expansions, the teeth can be displaced secondarily. In present case, biopsy, however, did not reveal the typical histologic "Chinese character" pattern encountered in fibrous dysplasia.[15]

Regional odontodysplasia

In regional odontodysplasia, the affected teeth may fail to erupt and exhibit a radiographic pattern described as pale, wispy tooth images with a lack of contrast between dentin and enamel. Soft tissue swelling also has been reported.[16-18] The histology in our case did not indicate regional odontodysplasia; however, the predominant tissue consisted of immature woven bone.

Gingival fibromatosis

Because of the maxillary enlargement and fibrous gingivae in the pre-maxillary area, this condition was briefly considered.[15] Review of the radiographs and histology was sufficient to rule out this condition.

Central hemangioma

Vascular lesions of this type are most often noted in patients between 10 and 20 years of age.[15] Radiographic imaging may show a honeycomb or soap-bubble appearance, and sometimes resorption of tooth roots in the area. Before biopsying the lesion, aspiration was attempted but no excessive blood flow occurred.

Tumor of bone

A variety of benign tumors that arise primarily within bone can be considered.[15] Included in this list are ossifying fibroma, cementifying fibroma, odontogenic myxoma, chondroma, developing odontoma, and calcifying epithelial odontogenic tumor. The results of the tissue examination along with the clinical and radiographic findings effectively ruled these lesions out.

Management

Treatment modalities for SOD are still insufficient. Due to the non-progressive nature and the non-severe form of facial asymmetry in SOD, recontouring osteotomy is not advised. Treatment can only be limited to restoring the functional dental occlusion in the affected side by orthodontic tooth movement and/or prosthetic replacement of missing teeth and should be delayed until the pubertal growth spurt has ended.[4,9]

In present case, surgical recontouring of pre-maxilla together with extraction of maxillary anterior permanent teeth was performed under general anesthesia. Patient was clinically asymptomatic after 6-month follow-up and was placed on interim prosthetic rehabilitation.

CONCLUSION

SOD is characterized by variability of its clinical and radiological features and may mimic other fibro-osseous lesions. Therefore, the definitive diagnosis of this lesion remains dependent on histological evaluation. In present case, gingivo-dento-alveolar pre-maxillary involvement was noticed which is first of its kind reported in world English literature. Therefore, such presentation should be considered while diagnosing a case of SOD.

Almost half of patients with SOD present with a facial cutaneous lesion which could be hypertrichotic, pigmentory, erythematous, commissural defect, or combination of these. This puts dermatologists in an ideal place to identify such patients, and as gingivo-dentoalveolar involvement is constant in all cases of SOD, the role of dentists is also crucial in early diagnosis.

Early recognition of SOD also requires appropriate referrals to medical colleagues and interdisciplinary team approach mandatory, so patients with SOD can gain access to specialist dental care, including orthodontics, prosthodontics, and oral surgery. Although SOD is a rare entity, clinicians should be aware of its presence when encountering patients presenting with maxillofacial asymmetry and should also consider it in differential diagnosis when treating a maxillofacial asymmetry unresponsive to treatment.

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